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Case Report

# Primary Hodgkin's disease of the thymus

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ABSTRACT

The thymus is a lymphoid organ located in the antero-superior mediastinum and composed of heterogeneous admixture of lymphoid and epithelial elements. Thymic tumors are dominated by thymoma; lymhomas are uncommon involving the gland secondary to adenopathies. Primary thymic lymphomas are exceptional.

We report a 25-year-old man referred to us because of chest pain and irritating cough since 2 months. Physical examination was normal and no lymph nodes were palpable. Chest radiography showed an anterior mediastinal mass. Computed tomography (CT) examination of the chest revealed a homogeneous tissular mass occupying the anterior mediastinum in its upper and medium stage, extending to the infundibulum without lymph node enlargement. Thymectomy and wedge were performed via a median sternotomy. Pathologic diagnosis was nodular sclerosis Hodgkin's disease of the thymus. The patient received 6 cycles of ABVD (Adriamycin [doxorubicin], bleomycin, vinblastin and dacarbazine) and mediastinal radiotherapy. Currently, tow years after diagnosis, he is well and free of disease.

Primary thymic Hodgkin's lymphoma is a rare entity which should be included in the differential diagnosis of anterior mediastinal mass especially in young patients.

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## 1. Introduction

The thymus is a lymphoid organ located in the antero-superior mediastinum and composed of heterogeneous admixture of lymphoid and epithelial elements. Thymic tumors are dominated by thymoma; lymhomas are uncommon involving the gland secondary to adenopathies. Primary thymic lymphomas are exceptional. We report a new case of Hodgkin thymic lymphoma without adenoapthies.

## 2. Case report

A 25-year-old man complained of chest pain and irritating cough for the past 2 months. Familial and personal histories were negative. At admission, he was in good general conditions. Physical examination was normal and no lymph nodes were palpable. Laboratory tests were within normal limits. Chest radiography showed an anterior mediastinal mass, eccentric to the right (Fig. 1). Computed tomography (CT) examination of the chest with infusion of contrast material revealed a homogeneous tissular mass occupying the anterior mediastinum in its upper and medium stage, extending to the infundibulum without lymph node enlargement (Fig. 2). Fiberoptic endoscopy was normal and cytological examination gave negative results. Computed tomographic scanning of the abdomen revealed no abnormalities. These findings were considered consistent with the diagnosis of thymoma, and surgical excision was undertaken through a median sternotomy. The mediastinum was explored: the tumor was developed effectively in the thymus. It was polylobulated, involving the mediastinal face of the right upper lobe and invading capsule. Biopsy of the tumor showed thymic tissue on frozen section examination. Thymectomy and wedge were performed after careful dissection from the pericard, phrenic nerves and great vessels. The patient had an uneventful postoperative recovery. Microscopic examination of the resected specimen revealed a thymic parenchyma infiltrated by nodules separated by fibrous septa. These nodules consisted of Hodgkin's cells, Reed–Sternenberg cells and lacunar cells intermingled with lymphocytes, plasmocytes, neutrophils and eosinophils. Lymph nodes next to mediastinal mass were not involved. Immunohistochemical study revealed that tumoral cells were CD15+, CD30+, CD20−, CD3−, EMA−. The histologic examination confirmed nodular sclerosis-type Hodgkin disease of thymic origin. Bone biopsy and abdomino-pelvien CT were negative. The tumor was classified stage IAaE. The patient received 6 cycles of ABVD (Adriamycin [doxorubicin], bleomycin, vinblastin and dacarbazine) and mediastinal radiotherapy. Currently, two years after diagnosis, he is well and free of disease.

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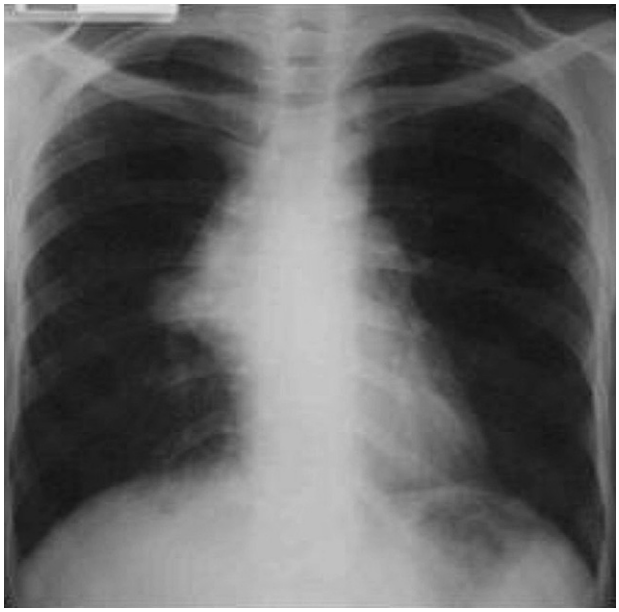

**Fig. 1.** Chest radiography: anterior mediastinal mass, eccentric to the right.

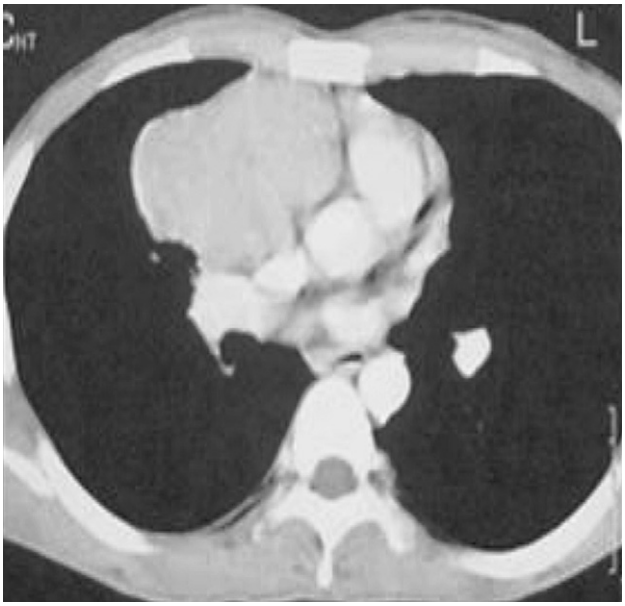


**Fig. 2.** Computed tomographic scanning of the chest: homogeneous tissular mass occupying the anterior mediastinum in its upper and medium stage, extending to the infundibulum without lymph node enlargement.

## 3. Comments

Hodgkin disease frequently involves intrathoracic structures, particularly mediastinal lymph nodes. Involvement of the thymus gland is known to occur; however, its exact prevalence is unknown.[1,2] Heron found 30% of thymic enlargement among 50 patients with evidence of thoracic disease on CT scans.[2] This frequency was higher (56%) in the series of Wernecke[1] because of the availability of follow up CT scans in all patients. However, Hodgkin disease involving the thymus gland without associated lymphadenopathy is rare.[3,4] In the series of Heron,[2] all cases with thymic enlargement at the diagnosis had also enlarged mediastinal lymph nodes. Among the 43 patients with newly diagnosed Hodgkin lymphoma, Wernecke[1] found only 6 cases with isolated thymic enlargement.

Thymus may be involved by malignant lymphoproliferative diseases, whether primarily or secondarily. Involvement by T-cell disorders such as lymphoblastic lymphoma is understood, but the pathogenesis of involvement by Hodgkin's disease remains unclear.[5,6]

Patients with thymic involvement are younger (second to third decades of life) than those without enlargement.[5] Most of the series showed male predominance.[3,7]

Clinically, patients with Hodgkin disease of thymic origin are asymptomatic until extrathymic disease develops.[4,8] On diagnosis, a third of the patients usually have fever, nocturnal sweating, and weight loss; chest symptoms being uncommon. However, in some cases, compression or invasion of mediastinal structures may trigger symptoms such as pain, coughing or dyspnea.[8]

Patients with thymic enlargement have generally radiological abnormalities in the form of an anterior mediastinal narrowing[8] as was true in our patient.

Patients with anterior mediastinal mass pose a diagnostic challenge. Differential diagnoses include lymphoma, thymoma, substernal thyroid mass, teratoma, germinal tumor and benign cyst of the thymus or simple hyperplasia.[9] CT has been shown to be the method of choice for evaluating mediastinal abnormalities offering information regarding the precise location, extent of the disease, relationship of the mass to other organs, and the response to the therapy.[9]

Thymic lymphoma appears as a discrete or lobulated mass which has typically homogeneous low attenuation. However, it can sometimes appear heterogeneous secondary to necrosis, hemorrhage, or cystic changes. Calcifications are rare, usually secondary to treatment.[10] In the series of Heron, it was easy to differentiate enlarged thymus from enlarged lymph nodes. In those patients with diffuse thymic enlargement, there was no significant difference in attenuation of the thymus compared with that of normal thymus. Since the density of thymus is similar to other mediastinal soft tissue structures in most patients, morphologic criteria must be used to identify the thymus. Both the position and shape of the thymus are characteristic.[3] On MR, the findings are similar to thymoma: low signal intensity on T1WI and high signal intensity on T2WI. In this last sequence, the tumor may show heterogeneity due to dense fibrosis (low signal intensity), edema, inflammation, and necrosis on cyst formation (high signal intensity).[10] In our case, there was no accompanying mediastinal disease, so the diagnosis of thymoma was initially suspected.

Surgical excision, thoracoscopy, mediastinoscopy, and biopsy thoracotomy are indicated in the approach of the anterior masses. Fine-needle aspiration has been used for the differential diagnosis of thymic lesion, especially when thymomas are suspected.[3] However, it is difficult to obtain a definitive preoperative diagnosis in most thymic lesions. Therefore, surgeons must often resort to an open biopsy, as was true in our case.

In thymic Hodgkin disease, the most common histologic type is nodular sclerosis. On histologic study, nodular sclerosis Hodgkin disease is characterized by a microscopic nodularity, composed of lymphoid cells interspersed with a marked inflammatory cell reaction and separated by wide fibrous bands. Identification of Reed–Sternberg cells confirms the diagnosis.[8]

## 4. Conclusion

Primary thymic Hodgkin's lymphoma is a rare entity which should be included in the differential diagnosis of anterior mediastinal mass especially in young patients.

## Conflict of interest

We confirm that none of authors have a conflict of interest to declare in relation to this work.

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